



Egyptian Society of Ear, Nose, Throat and Allied Sciences  
**Egyptian Journal of Ear, Nose, Throat and Allied Sciences**

[www.ejentas.com](http://www.ejentas.com)



## CASE REPORT

# Fibrosarcoma of the nasal cavity: A case report



Omar Maliki <sup>a,\*</sup>, Oleksandr Aleksandrov <sup>a</sup>, Phillippes Carles <sup>a</sup>, Xavier Planquart <sup>a</sup>,  
 Emmanuelle Vaz <sup>b</sup>, Catherine Bertrand <sup>b</sup>, Andrzej Rosikon <sup>a</sup>

<sup>a</sup> Department of Otorhinolaryngology and Head and Neck Surgery, Robert Ballanger Hospital, Aulnay, France

<sup>b</sup> Department of Pathology, Robert Ballanger Hospital, Aulnay, France

Received 31 January 2014; accepted 4 April 2014

Available online 29 April 2014

### KEYWORDS

Fibrosarcoma;  
 Nasal cavity;  
 Sudden death

**Abstract** Nasal fibrosarcoma is an infrequent malignant neoplasm. It usually presents as other sarcomas in this region, with nasal obstruction and epistaxis. The final diagnosis is based on the histopathology and immunohistochemistry. We report the case of a 37-year-old man with a 3-month history of recurrent epistaxis and nasal obstruction. Nasal endoscopy confirmed a right nasal neoplasia. Computed tomography and magnetic resonance image showed the tumor. TEP scan showed no metastasis. Complete removal was achieved through a combined surgery, by endoscopic endonasal approach and by incision in the right upper oral vestibule. Fibrosarcoma was found on histopathologic and immunohistochemistic examinations. After 12 months, the postoperative course was uneventful and follow-up information showed no recurrence of metastasis. However, in the 13th month, the patient suddenly died at home. Autopsy found no obvious cause for his death. To the best of our knowledge, no case of a fibrosarcoma of the nasal cavity with sudden death has been previously reported in the English-language.

© 2014 Egyptian Society of Ear, Nose, Throat and Allied Sciences. Production and hosting by Elsevier B.V. All rights reserved.

## 1. Introduction

Nasal fibrosarcoma is an infrequent malignant neoplasm.<sup>1</sup> These tumors only account for 7–10% of all the sarcomas of the head and neck region.<sup>2</sup> Only a few scattered cases have been published. The etiology of fibrosarcoma remains obscure although radiation exposure has been thought to be the most important etiological factor, followed by trauma, and underlying conditions of bone like Paget's disease, fibrous dysplasia,

or chronic osteomyelitis. The diagnosis is based on histopathology and immunohistochemistry.

The treatment of choice is radical surgery. Radiation therapy and chemotherapy<sup>3</sup> can be used in inoperable cases. Prognosis of the tumor is dependent on histological grade, tumor size and adequate surgical treatment with disease free margins.<sup>3</sup> We report a patient with nasal fibrosarcoma, which was successfully managed by endoscopic endonasal and oral surgery who faced an unexplained sudden death in the 13th month after surgery.

## 2. Case report

A 37-year-old man patient was referred to our hospital with 3 months history of enlarging mass in the right nasal cavity,

\* Corresponding author. Tel.: +33 1 49 36 70 99; fax: +33 1 49 36 70 75.

E-mail address: [dr.omar.ent@gmail.com](mailto:dr.omar.ent@gmail.com) (O. Maliki).

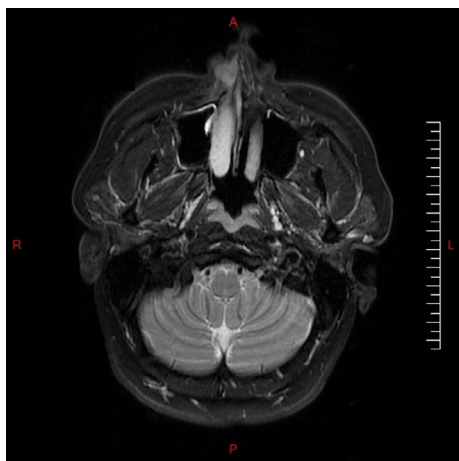
Peer review under responsibility of Egyptian Society of Ear, Nose, Throat and Allied Sciences.

intermittent epistaxis and right nasal obstruction. Medical history was peptic ulcer. Endoscopic examination revealed a hemorrhagic black mass completely filling the anterior part of the right cavity extending outwards from the vestibule. No other pathology was detected from the nasopharynx, oral cavity and pharyngo-larynx. There were no palpable lymph nodes in the neck. Routine hematological and biochemical tests were normal. Computed tomography (CT) scans of the nose and sinuses revealed an iso-dense mass filling the anterior part of the nasal cavity without bony erosion. The paranasal sinuses, orbita and cranial fossa were uninvolved. The mass exhibited a low-intensity signal on a T1-weighted magnetic resonance image (MRI) and a high-intensity signal on a gadolinium-enhanced T1-weighted image and a T2-weighted image, measuring 25 × 14 mm (Fig. 1). A biopsy was performed under local anesthesia and the histopathologic examination reported an unencapsulated spindle cell tumor with a herringbone pattern without epithelial differentiation. The neoplastic cells were homogeneous with little pleomorphism and little cytoplasm, but containing hyperchromatic nuclei. Mitoses were seen at a frequency of 7/10 high power fields. A variable amount of collagen was observed (Fig. 2). Immunohistochemistry showed the tumor to stain diffusely for CD99 and vimentin. Cytokeratins, EMA, actin, desmin, melanin A, S-100, CD33, CD34, P63 and bcl.2 were negative. There was a moderate to focally high Ki-67 staining of 50%. Abdominal and chest CT did not show any evidence of metastases. PET scan showed an isolated hypermetabolic localization (SUV = 33,84) in the floor of the right nasal cavity without any secondary lesion (Fig. 3). Under general anesthesia, the tumor was completely resected due to a combined surgical approach, by nasal and by right upper oral vestibule (Fig. 4).

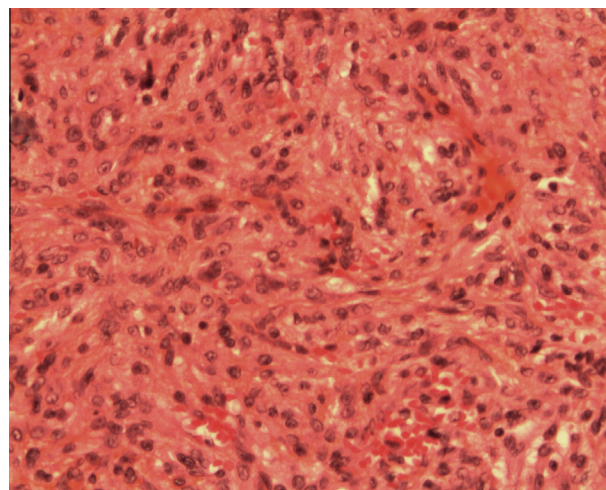
The postoperative course of the patient was uneventful and follow-up information showed no recurrence of metastasis after 12 months. However, in the 13th month, the patient suddenly died at home. Autopsy found no obvious cause for his death.

### 3. Discussion

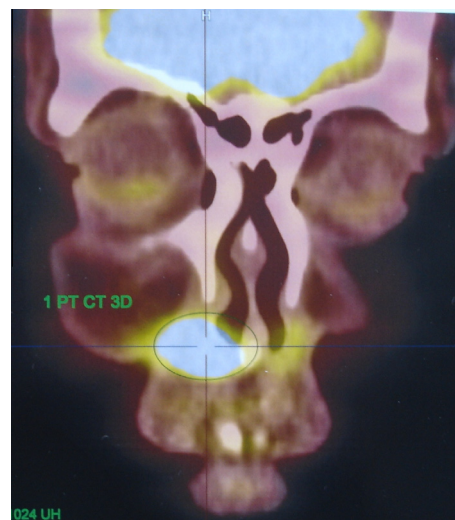
Fibrosarcomas are a malignant tumor of fibroblasts. The localization of the head and neck is uncommon. Approxi-



**Figure 1** Axial MR T2-weighted showing a well-defined, high-intensity tumor in the right nasal cavity.



**Figure 2** This is a low-grade sarcoma composed of spindle cells with little pleomorphism but with hyperchromatic nuclei and moderate mitotic activity, arranged in fascicles and bundles with occasional "herringbone" pattern (hematoxylin and eosin stain × 200). Immunohistochemically, the cells are only positive for vimentin and CD99 (not shown).

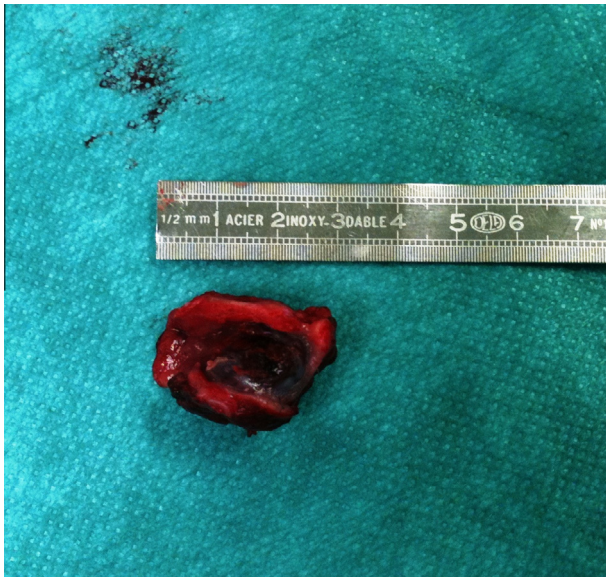


**Figure 3** PET scan showing an isolated hypermetabolic localization in the floor of the right nasal cavity.

mately 19% of all soft-tissue sarcomas are fibrosarcomas, and only 16% of these occur in the head and neck.<sup>4</sup> Only a few cases have been published, but there are several comprehensive reviews on the histopathologic findings and treatment results.

It usually presents in the third and fourth decades of life, more commonly among males.<sup>2</sup>

The exact cause of fibrosarcoma is not entirely understood. However, studies have indicated that genetic alterations may play a role. A chromosomal rearrangement has been found in some fibrosarcomas. Radiotherapy to the local site has been proposed as a predisposing factor for increased risk of fibrosarcoma. Disease processes like Paget's disease and osteomy-



**Figure 4** Tumor after resection.

elitis have also been implicated in few cases in which the fibrosarcomas developed in the bone.<sup>3</sup>

These neoplasms appear to originate in the periosteum rather than in mucosal connective tissue.<sup>5</sup> Fibrosarcomas are usually unencapsulated, slowly growing spindle cell tumors.<sup>6</sup> They may invade or atrophy adjacent bone by direct pressure.<sup>5</sup> Nasal fibrosarcoma demonstrates variable collagen production, does not show differentiation in other types of tissue, and can metastasize.<sup>1</sup> These features allow establishment of a differential diagnosis between inflammatory myofibroblastic tumor, fibroma, desmoid fibromatosis, and nasal fibrosarcoma. Other histological types have been described and can lend confusion as synovial sarcoma and Ewing's sarcoma. Histologically, nasal fibrosarcomas are hypercellular lesions composed of thin, elongated spindle cells arranged in long fascicles and bundles intersecting at different angles (the herringbone pattern) with a great degree of pleomorphism and nuclear atypia and large number of mitoses, and may have areas of prominent necrosis. Immunostaining for CD99 and vimentin is positive. The tumor is negative for desmin, actin and S-100 protein.

Despite the paucity of cases of nasal fibrosarcoma in the literature, the initial treatment of choice appears to be a wide local excision. Endoscopic removal of nasal malignant tumors has been thoroughly reported, with good oncologic results and less morbidity than radical approaches.<sup>7</sup>

Radiotherapy has been used as an adjuvant treatment for incomplete gross excision or positive microscopic margins. The role of chemotherapy as a sole treatment for the head and neck fibrosarcoma warrants further study. Chemotherapy has been employed as a neoadjuvant treatment in combination

with radiotherapy as a palliative measure for patients with unresectable tumors.<sup>2</sup>

Nasal fibrosarcoma is usually associated with a high risk of local recurrence, and low risk of distant metastases.<sup>3</sup> Frequent endoscopic and imaging controls are very useful to achieve early detection of recurrences that can also be treated endoscopically.<sup>8</sup>

Prognosis of the tumor is dependent on histological grade, tumor size and adequate surgical treatment with disease free margins. The 5-year survival rate for this disease is poor, ranging from 20% to 35%.<sup>3</sup>

To the best of our knowledge, our patient represents the first case of nasal fibrosarcoma who died of sudden death, without metastasis, that has been reported in the English-language literature. Sotolongo Vergo in Cuba report a case of sudden death in a retroperitoneal fibrosarcoma excised 4 month before and necropsy concluded to a right pulmonary thromboembolism, abdominal ganglionic metastases and retroperitoneal tumor extension.<sup>9</sup> Until now, we have no explanations for the death of our patient who have no medical history particularly cardiac. We do not know how far we can relate this death to his nasal fibrosarcoma.

#### Conflict of interest

None.

#### References

1. Perez-Ordóñez B, Huvos AG. Nonsquamous lesions of nasal cavity, paranasal sinuses, and nasopharynx. In: Gnepp DR, ed. *Diagnostic Surgical Pathology of the Head and Neck*. Philadelphia: W.B. Saunders Company; 2001:79–89.
2. Koka V, Vericel R, Lartigau E, et al. Sarcomas of nasal cavity and paranasal sinuses: chondrosarcoma, osteosarcoma and fibrosarcoma. *J Laryngol Otol*. 1994;108(11):947–953.
3. Wadhwan V, Chaudhary MS, Gawande M. Fibrosarcoma of the oral cavity. *Indian J Dent Res*. 2010;21:295–298.
4. Batsakis JG, Rice DH, Howard DR. The pathology of head and neck tumors: spindle cell lesions (sarcomatoid carcinomas, nodular fasciitis, and fibrosarcoma) of the aerodigestive tracts, Part 14. *Head Neck Surg*. 1982;4(6):499–513.
5. Alameda YA, Perez-Mitchell C, Busquets JM. Nasal cavity ossifying fibrosarcoma: an unusual fibro-osseous neoplasm. *Ear Nose Throat J*. 2010;89(11):E1–3.
6. Heffner DK, Gnepp DR. Sinonasal fibrosarcomas, malignant schwannomas, and "Triton" tumors. A clinicopathologic study of 67 cases. *Cancer*. 1992;70(5):1089–1101.
7. Bercin S, Muderris T, Kırıs M, et al. A rare sinonasal neoplasm: fibrosarcoma. *Ear Nose Throat J*. 2011;90(5):E6–8.
8. Plaza G, Ferrando J, Pinedo F. Sinonasal fibrosarcoma: a case report. *Eur Arch Otorhinolaryngol*. 2006;263(7):641–643.
9. Sotolongo Vergo I, Suárez Cabrera RE, Arencibia García J. Retroperitoneal fibrosarcoma. A propos of a case. *Arch Esp Urol*. 2003;56(4):421–424.